of renal tubular epithelial cells. Several of the cellular and molecular mechanisms involved in cystogenesis have been identified.

Renal cysts can be acquired, hereditary, or developmental. Acquired conditions include cystic kidney disease that develops in patients with existing ESRD. Cystic kidney diseases also are important causes of ESRD.

The most common inherited cystic kidney diseases are the polycystic kidney diseases (PKDs), including autosomal dominant and autosomal recessive forms of PKD. Other hereditary cystic renal diseases include medullary cystic kidney disease (MCKD), Von Hippel–Lindau disease (VHLD), and tuberous sclerosis. Developmental cystic diseases of the adult kidney include localized renal cystic disease, multicystic dysplastic kidney, and medullary sponge kidney.

In the inherited disorders, several mutations have been associated with cyst formation. Mutation of any of the tubular epithelial–related genes such as *PKD1*, *PKD2*, *MUC1* (formerly *MCKD1*), and *MCKD2* can result in disruption of normal ciliary function, resulting in cyst formation from overproliferation of tubular epithelium and increased fluid secretion. In PKD, the cysts are not connected to the urinary drainage system, and cellular secretion results in cyst enlargement.

Simple Cysts

Widespread use of ultrasonography and CT has resulted in frequent detection of kidney cysts. Simple cysts are most common. They are usually unilateral, solitary, well-defined structures, but they can be multiple and bilateral. They tend to be more common among older adults and are often benign, incidental findings on radiographic imaging. Sonography reveals a thin-walled, fluid-filled cavity with no septations or calcifications. The diameter varies between 0.5 and 1.0 cm, but a few may be as large as 3 to 4 cm in diameter.

Simple cysts are usually asymptomatic but occasionally may result in a palpable abdominal mass, infection, back pain, or hematuria. Differentiation of simple cysts from cysts associated with genetic disorders is based on the cystic pattern, age at detection, and family history.

In the absence of symptoms, no treatment is required for simple cysts. If the kidney cyst becomes infected, causes pain, or leads to renin-mediated hypertension, percutaneous drainage is often the first step in further evaluation and management.

Complex Cysts

Differentiation of simple from complex cysts is usually made radiographically. When in doubt, histologic examination is required to exclude malignancy, but imaging is sensitive and specific, and it suffices in most cases. The distinction between complex and simple cysts is important in monitoring the need for intervention because simple cysts are usually benign, whereas complex cysts have a higher risk of malignancy and other complications.

Initial evaluation includes ultrasonography and triphasic CT to characterize the cyst. If the cyst characteristics of size, nodularity, mural enhancement, or septations change over time, the likelihood of malignancy increases. In a simple cyst, complications resulting from hemorrhage or infection can result in the features of more complex cysts, including calcification, septa, irregular borders, and multilobularity.

To help with diagnosis and management, the Bosniak classification of renal cyst was introduced in 1996 and revised in 2003. This classification, which includes four categories based on triphasic CT findings, is described in Table 29-5. Category 1 cysts are benign, and category II cysts have a 0% to 5% risk of malignancy. The risk increases to almost 50% for category III cysts. There are also several important subcategories. Category III and IV renal cysts are considered to be renal carcinoma unless proved otherwise, and they should be surgically resected.

POLYCYSTIC KIDNEY DISEASE

Autosomal dominant polycystic kidney disease (ADPKD) is a common cause of cystic renal disease and an important cause of ESRD. The monogenetic, progressive disorder is characterized by multiple cysts in kidneys and other organs, including the liver and pancreas. The incidence of ADPKD is 1 case in 400 to 1000 live births, and between 300,000 and 600,000 Americans are affected by the disease.

Mutations in the *PKD1* and *PKD2* genes are responsible for about 85% and 15% of ADPKD cases, respectively, and there is evidence for important modifier genes. *PKD1* is located on chromosome 14 and encodes the protein polycystin 1 (PC1), which functions as a membrane receptor. *PKD2* is located on chromosome 4 and encodes polycystin 2 (PC2), which functions as a calcium permeable cation channel (E-Fig 29-4). PC1 and PC2 regulate intracellular calcium homeostasis and signaling pathways involved in tubular morphogenesis and cell-cell

TABLE 29-5 BOSNIAK RENAL CYST CLASSIFICATION SCHEME

CATEGORY	DESCRIPTION
I. Simple cyst	A benign simple cyst with a thin wall and no septa, calcifications, or solid components.
II. Minimally complicated	A benign cystic lesion with a few thin septa. The wall or septa may contain fine calcifications or short segment of a slightly thickened calcification. (This category also includes uniformly high-attenuating lesions that are less than 3 cm in diameter, well marginated, and nonenhancing.)
IIF. Complicated	Well-marginated cysts but more complicated than category II. They have multiple thin septa or minimal smooth thickening of the septa or wall and may contain calcifications that may be thick and nodular. (This category also includes totally intrarenal, nonenhancing high-attenuating lesions that are more than 3 cm in diameter.)
III. Indeterminate	Indeterminate cystic masses that have thickened, irregular, or smooth walls or septa. These lesions are enhancing on computed tomography. Between 40% and 60% of lesions are malignant (e.g., cystic renal cell carcinoma, multiloculated cystic renal cell carcinoma). The remaining lesions are hemorrhagic, chronic, infected cysts or multiloculated cystic nephroma and are benign.
IV. Malignancy	On computed tomography, they have characteristics of category III cysts and contain enhancing soft tissue components that are adjacent to and independent of the wall or septum on the cyst. Between 85% and 100% of lesions are malignant; evaluation and surgical excision are recommended.